

# Lower leg pain, fever and swelling

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#### **DIFFERENTIAL DIAGNOSIS**



#### **PEAK AGE**

 0-5 yrs : LCH, metastatic neuroblastoma, leukemia

• 5-10 yrs : Osteosarcoma

• 10-20 yrs : Ewing's Sarcoma





















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R

ANTERIOR



LT LAT RT MED



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#### **EWINGS' SARCOMA**

- **Neuroectodermal** origin.
- Common Karyotype translocation long arms of chromosome 11 and 22.
- Ewing sarcoma, PNET, and Askin tumor cytogenetically closely related lesions on a morphologic continuum, which is referred to as the **Ewing sarcoma family of tumors**.
- 2<sup>nd</sup> most common primary malignant tumor; 1<sup>st</sup> osteosarcoma.
- Slight male predominance; peak age of occurrence is 10-15 years.
- Clinical presentation of Ewing sarcoma is usually nonspecific, with **pain, mass, swelling, fever.**

#### IMAGING CHARACTERISTICS

#### Widespread Skeletal distribution :

Femur Ilium Tibia Humerus Fibula Ribs Sarcoma

Majority of the lesions are actually **meta-diaphyseal**; not diaphyseal as typically described !



### RADIOGRAPHY AND CT

Overwhelming majority : Bone destruction with a **moth-eaten** to **permeative pattern** and a wide zone of transition.

Hair on end aggressive periosteal reaction or "Onion Peel"; better appreciated on Plain film radiography and CT.

Linear low attenuation channels extending through the cortex, allowing continuity between the medullary cavity and soft tissue mass.

Saucerization or extrinsic erosion of the bone outer cortex is more often seen in cases extending from the medullary canal.

### MRI

- T1: low to intermediate signal.
- T2: heterogeneously high signal, may see hair on end low signal striations.
- T1 C+ (Gd): heterogeneous enhancement.
- Extra-skeletal tumor: **Detection of high-flow vessels** within the lesion at MR imaging, while not specific, is an additional finding suggestive of this diagnosis.

# NUCLEAR MEDICINE

Increased uptake on bone scintigraphy.

Markedly FDG avid : treatment response.

Maximum SUV of less than 2.5 after chemotherapy associated with increased progression-free survival and a positive predictive value for favorable response.

# **TREATMENT & PROGNOSIS**

- Multi disciplinary management.
- Neo adjuvant and adjuvant chemotherapy.
- Goal of surgical resection is to maximize local control.
- Surgical treatment as the primary therapy for local control requires complete resection with wide or radical margins.

#### References

- From the Radiologic Pathology Archives: Ewing Sarcoma Family of Tumors: Radiologic-Pathologic Correlation Mark D. Murphey, MD, Mark D. Murphey, Lien T. Senchak, MD, Lien T. Senchak, Pramod K. Mambalam, MD, Pramod K. Mambalam, Chika I. Logie, MD, Chika I. Logie, Mary K. Klassen-Fischer, MD, and Mary K. Klassen-Fischer, Mark J. Kransdorf, MD, Mark J. Kransdorf.
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